Apical Hypertrophic Cardiomyopathy with Severe Myocardial Bridging in a Syncopal Patient

Shih-Chung Chen, Kuang-Te Wang[1], Charles Jia-Yin Hou, Yu-San Chou and Cheng-Ho Tsai

A 48-year-old male presented to the emergency department with the manifestation of syncope on exercise. Electrocardiography showed giant negative T wave in V3-V6. Transthoracic echocardiography revealed apical hypertrophic cardiomyopathy (AHC), trivial mitral regurgitation and no pressure gradient across the left ventricular outflow tract. His coronary angiogram demonstrated normal coronary arteries and myocardial bridging in the middle portion of the left anterior descending artery causing severe vessel squeezing. Another episode of syncope developed at the postural change during admission. The mechanism of his syncope may be associated with hemodynamic instability related to a myocardial bridging. Increased Left ventricle (LV) contractility or decreased preload will exacerbate vessel squeezing in patients of apical hypertrophic cardiomyopathy with severe myocardial bridging. Our patient was treated with propranolol orally 30 mg per day and was advised to avoid strenuous exercise. He has remained free of syncope to date.

Key Words: Syncope • apical hypertrophic cardiomyopathy • myocardial bridging

INTRODUCTION

Myocardial bridging has been reported with a prevalence of 0.5-16% in coronary angiograms.[1] It is characterized by a systolic narrowing of a coronary artery, mainly the left anterior descending coronary artery, responsible for a milking effect.[2] It can be associated with a variety of cardiac diseases, particularly hypertrophic cardiomyopathy (HCM). Apical hypertrophic cardiomypathy (AHC) is more common in Asian population.[3] However, the coexistence of AHC with myocardial bridging is extremely rare.[4] Although myocardial bridging is generally considered benign, it may be associated with myocardial ischemia, infarction, ventricular arrthymia and sudden death.[5-7] Syncope is uncommon in AHC. The mechanisms of syncope in AHC remain controversial.

We describe a patient with AHC and severe myocardial bridging of the left coronary artery who developed syncope that was resolved with beta-blocker.

CASE REPORT

A 48-year-old male presented to the emergency department with the manifestation of syncope during a badminton play. Dizziness was encountered at the play. The syncope developed without any prodromes. He had no other cardiovascular symptoms and no family history of cardiovascular disease. His heart rate was 78 beats per minute, blood pressure was 131/78 mmHg, respiratory rate was 19/min and the temperature was 36 °C. Physical examination showed negative findings except a left-displaced apical impulse. The remainder of the physical examination produced normal finding. The resting electrocardiogram (ECG) showed first-degree atrioventricular block, right atrial enlargement, incomplete right bundle

Received: March 12, 2002 Accepted: March 3, 2003
Division of Cardiology, Department of Internal Medicine, Mackay Memorial Hospital [1]Taitung Branch and Taipei, Taiwan.
Address correspondence and reprint requests to: Dr. Kuang-Te Wang, Division of Cardiology, Department of Internal Medicine, Taitung Mackay Memorial Hospital Taitung Branch, No. 1, Lane 303, Chang Sha Street, Taitung 950, Taiwan. Tel: 886-89-310150.

branch block and giant negative T wave in V3- V6 (Figure 1). Measurement of cardiac enzyme yielded normal values. The transthoracic echocardiography demonstrated left ventricular hypertrophy confined to the apical region, trivial mitral regurgitation and preserved left ventricular contractility with ejection fraction of 67%. The ratio of apical to basal ventricular septal thickness was 1.56. The continuous-wave Doppler pressure gradient over the left ventricular outflow tract (LVOT) was 5.5 mmHg at rest. AHC was diagnosed. After echo- cardiographic evaluation, the patient felt nausea and presented with another episode of syncope at postural change, accompanying by a BP drop. He regained consciousness without any resuscitation 30 seconds later.

The 24-hr Holter monitor demonstrated persistent ST depression and giant T wave inversion. No supra-ventricular or ventricular arrthymia was identified. Tilt table test was performed to evaluate the possibility of vasovagal syncope. When the table was lifted to 70 degrees, he complained of dizziness in association with horizontal ST depression identified by ECG reading. No orthostatic hypotension or arrthymia was documented. Cardiac catheterization was performed and showed a myocardial bridge at the middle portion of the left anterior descending coronary artery and septal perforators (Figure 2A). The coronary arteries were otherwise normal (Figure 2B). Left ventriculography showed normal left ventricular contractility, no regional wall motion abnormality, mild mitral regurgitation and spadelike configuration of the left ventricle (Figure 4). Cardiac catheterization recorded a pressure gradient of 6 mmHg across the LVOT. One mg of nitroglycerin was adminis-trated intracoronarly and augmented the angiographic appearance of the myocardial bridge (Figure 3). The LVOT pressure gradient increased from 6 mmHg to 15 mmHg after nitroglycerin injection. The mean arterial pressure decreased from 103 mmHg to 72 mmHg. The ECG recording showed dynamic ST change. The patient complained of dizziness after nitroglycerin injection. Then he was treated with beta-blocker (propranolol 10 mg tid) and remained free of syncope. Despite our recommendation, however, he only took the medication for 10 months.

Twelve months later, 12-lead ECG showed no giant

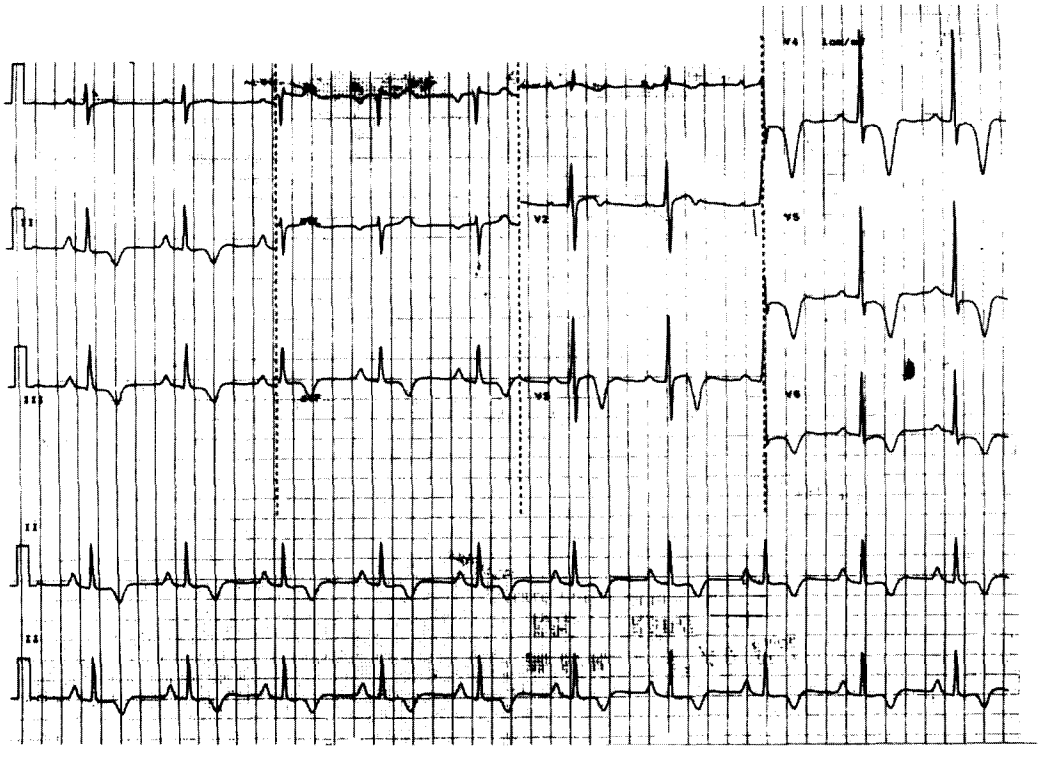

Figure 1. *Electrocardiography showing prominent T wave inversion in V3-V6. T wave pattern was caused here by hypertrophic cardiomyopathy. The echocardiography in this patient showed concentric left ventricular hypertrophy.*

negative T wave. However, follow-up treadmill exercise test demonstrated 2 mm horizontal ST depression at II, III, aVF and V4-V6 during stage 2 of Bruce protocol, accomparined with the symptom of dizziness. Immediate transthoracic echocardiography revealed a pressure gradient of 8 mmHg across the LVOT. At peak exercise, systolic arterial pressure was 145 mmHg, compared to 130 mmHg at rest.

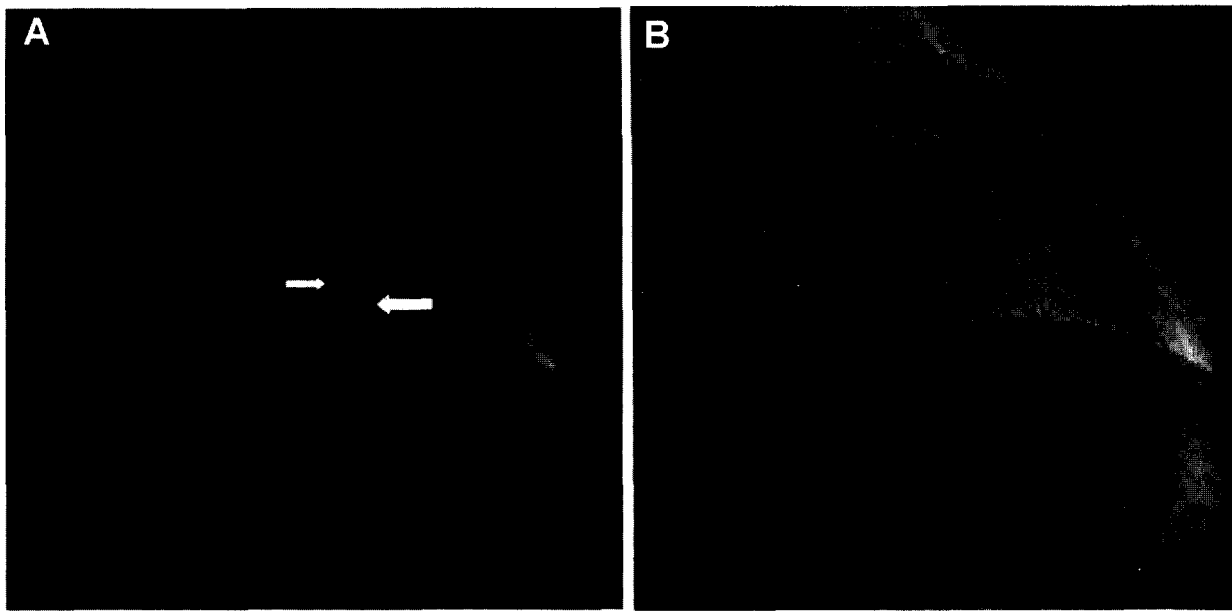

Figure 2. *(A) Angiographic appearances of the left anterior descending coronary artery demonstrated the characteristic sawfish appearance of the striking myocardial bridging of 20 mm length. Stenosis of the middle portion of LAD (large arrow) in systole was 70%. The septal perforators (small arrow) was totally compressed by hypertrophic cardiomyopthy in systole. (**B**) LAD appeared normal during diastole.*

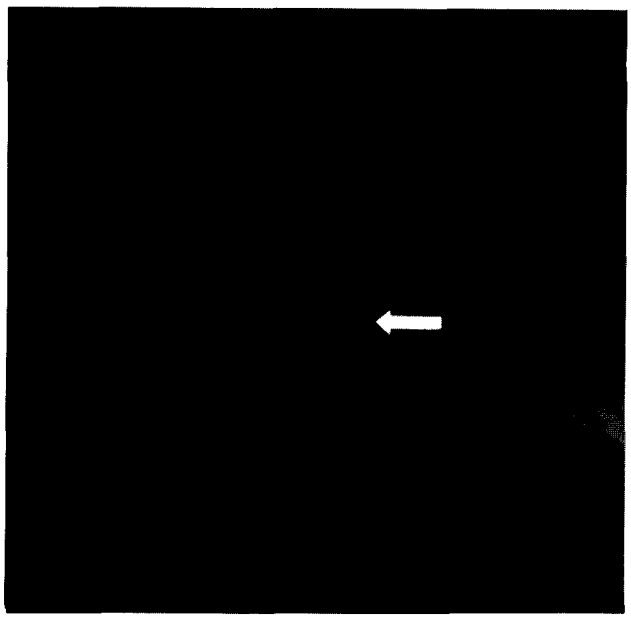

Figure 3. *After administration of nitroglycerin, coronary arteries including myocardial bridging dilated in diastole. Myocardial bridging was augmented as observed by angiography in systole. Stenosis was 87 % in systole. LVOT gradient = 40 mmHg .*

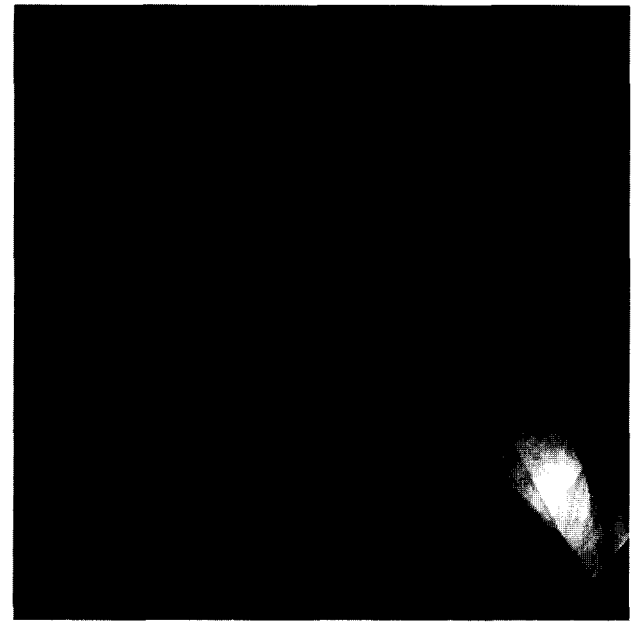

Figure 4. *Left ventriculogram showed ace of spades configuration in the right anterior oblique projection during end diastole.*

DISCUSSION

AHC is a variant of HCM with predominant involvement of the apex. The typical features include giant negative T wave in the precordial ECG leads, ace of spades configuration during angiographic imaging of the left ventricle in right anterior oblique projection and the absence of pressure gradient in the LVOT.[8] In Japan, AHC constitutes 25% of HCM. Beyond Japan, only 1% to 2% of cases are associated with AHC. AHC is considered to have negative family history. Also, it carries favorable prognosis when compared with the other form of HCM, which has an annual mortality of 2 to 3%.[9] From the findings of ECG, echocardiography and coronary angiography, our patient showed the typical picture of AHC.

Syncope is a characteristic symptom of HCM and is often provoked by exertion or postural change. An abnormal exercise blood pressure response, usually a failure of blood pressure to rise with exercise, was observed in 58% of HCM patients with syncope. However, syncope is uncommon in nonobstructive HCM, including AHC.[5]

The mechanisms of syncope in HMC involved multiple factors, including increasing left ventricular obstruction on exertion, altered diastolic filling, myocardial ischemia and abnormal systemic vascular tone in nonexercising muscle beds. Yoshida *et al.* demonstrated exercise-induced abnormal blood pressure response in patients with HCM was related to subendocardial ischemia during exercise.[10] Our patient experienced his first episode of syncope during exercise. He had abnormal blood pressure response during treadmill exercise test and complained of dizziness. Since no increase of pressure gradient across LVOT was documented, it is assumed that increase contractility during exercise may exacerbate vessel squeezing, trigger myocardial ischemia and cause abnormal blood pressure response and syncope in our patient. In addition, decreased preload, e.g. after administration of intracoronary nitroglycerin and during tilt table test, may exacerbate myocardial bridging and myocardial ischemia and may also contribute to his syncope.

Myocardial bridging is marked by systolic compression of an epicardial coronary arterial segment by the overlying myocardium. Vascular compression may persist into early and middiastole, causing reduction in coronary flow reserve. The common denominator for most symptomatic patients is a degree of systolic compression of 75% or more. Bridging can cause myocardial ischemia, infarction, ventricular arrthymia and sudden death. Myocardial bridging with compression of an epicardial coronary artery occurs in 30 to 50 percent of adults who have HCM.[11] Nevertheless, AHC with myocardial bridge was a rare entity.[4]

The treatment of myocardial bridging is based primarily on a pharmacological intervention. Beta-blockers remain the first-line therapy and may reduce the degree of systolic coronary artery narrowing, increase coronary vascular tone and lengthen diastole. However, not all patients achieve persistent symptomatic relief.[12,13] The other therapeutic approaches consist of surgical intervention (either surgical myotomy or bypass grafting). Surgical myotomy has been considered the treatment of choice for patients with medical failure. Nevertheless, deep incision of the ventricular wall is often needed. It carries the risk of mural aneurysm and scar formation with subsequent recurrent myocardial squeezing.

Recently, intracoronary stent implantation was been suggested to be an optimal treatment for the patients with myocardial bridging, allowing internal stabilization of the coronary artery lumen against external compression. Klues *et al.* reported immediate normalization of the flow velocity profile, lumen diameters, intracoronary pressures, and coronary flow reserve in 3 patients with stent deployment.[14] Haager *et al.* found that lumen diameters did not change significantly between 7 weeks and 6 months of follow-up in 11 patients with symptomatic myocardial bridging.[15] Hemodynamic alteration caused by mural compression can be neutralized by stent delivery.

In conclusion, we present herein a case of AHC with myocardial bridge and clinical manifestation of syncope. The underlying mechanisms remained elusive, but may involve increased myocardial contractility and decreased preload, which may exacerbate myocardial bridging. The patient responded well to beta-blocker.

REFERENCES

1. Angelini P, Trivellato M, Donis J. Myocardial bridges: a review. *Prog Cardiovasc Dis* 1983;26:75-88.
2. Portsmann W, Iwig J. Die intramurale Koronarie in Angiogramm. *Fortschr Roentgenstr* 1960;92:129-32.

3. Ko YL, Lei MH, Chiang FT, et al. Apical hypertrophic cardiomyopathy of the Japanese type: occurrence with familial hypertrophic cardiomyopathy in a family. *Am Heart J* 1992;124: 1626-30.
4. Partanen J, Kupari M, Heikkila J, et al. Left ventricular aneurysm associated with apical hypertrophic cardiomyopathy. *Clin Cardiol* 1991;14:936-9.
5. Juilliere Y, Berder V, Suty-Selton C, et al. Isolated myocardial bridges with angiographic milking of the left anterior descending coronary artery: a long-term follow-up study. *Am Heart J* 1995;129:663-5.
6. Endo M, Lee YW. Hayashi H, et al. Angiographic evidence of myocardial squeezing accompanying tachyarrhythmias as a possible cause of myocardial infarction. *Chest* 1978;73:431-3.
7. Morales AR, Romanelli R, Boucek RJ. The mural left anterior descending coronary artery, strenuous exercise and sudden death. *Circulation* 1980;62:230-7.
8. Smolders W, Rademakers F, Conraads V, et al. Apical hypertrophic cardiomyopathy. *Acta Cardiol* 1993;48:369-83.
9. Reddy V, Korcarz C, Weinert L, et al. Apical hypertrophic cardiomyopathy. *Circulation* 1998;98:2354.
10. Yoshida N, Ikeda H, Wada T, et al. Exercise-induced abnormal blood pressure responses are related to subendocardial ischemia in hypertrophic cardiomyopathy. *J Am Coll Cardiol* 1998;32:1938-42.
11. Mckenna W, Deanfield J, Faruqui A, et al. Prognosis in hypertrophic cardiomyopathy: role of age and clinical, electrocardiographic and hemodynamic features. *Am J Cardiol* 1981; 47:532-8.
12. Mazzu A, Di Tano G, Cogode R. Myocardial bridging involving more than one site of the left anterior descending coronary artery: an uncommon cause of acute ischemic syndrome. *Cathet Cardiovasc Diagn* 1995;34:329-32.
13. Nair CK, Dang B, Heintz MH. Myocardial bridges: effect of propranolol on systolic compression. *Can J Cardiol* 1986;2:218-21.
14. Klues HG, Schwarz ER, vom Dahl J, et al. Disturbed intracoronary hemodynamics in myocardial bridging: early normalization by intracoronary stent placement. *Circulation* 1997;96:2905-13.
15. Haager PK, Schwarz ER, vom Dahl J, et al. Long term angiographic and clinical follow up in patients with stent implantation for symptomatic myocardial bridging. *Heart* 2000; 84:403-8.

心尖肥厚性心肌症伴隨嚴重冠狀動脈壓迫症導致暈厥之探討

陳識中　王光德　侯嘉殷　周友三　蔡正河
台北市　馬偕醫院 心臟內科

一位四十八歲男性於打羽毛球時發生暈厥的現象,而被送至急診求治,心電圖顯示於胸前導程有巨大負向 T 波,胸部心臟超音波發現此病人有心尖心肌肥厚,輕度僧帽瓣逆流。

此外病患的冠狀動脈攝影顯示出於左前降支冠狀動脈有嚴重的血管壓迫症,左心室攝影有典型黑桃紙牌狀外觀。

另一次的暈厥發生於住院中,病患起身坐於輪椅上,有噁心的先前症狀。

此病患暈厥的機轉與血管壓迫症有關,運動與姿勢變化皆會加重血管壓迫,導致心肌缺氧,造成不正常血液動力學變化。病人以 propranolol 每天 30mg 治療,並提醒患者避免激烈運動,與突發性的姿勢變換，經過十二個月臨床追蹤,至今仍無復發現象。

關鍵詞：暈厥、心尖肥厚性心肌症、冠狀動脈壓迫症。